respectively. The overall median survival from stereotactic radiotherapy was 24.2 months and one patient was alive and disease free at 53 months. Based on these results, the authors suggested that some of these patients may have a prolonged survival or even may be cured.[7]

The two patients treated with CyberKnife radiosurgery remain stable at short-term follow-up. Longer follow up is needed to determine if this modality is beneficial to patients who harbor such disease. Based on previous reports that have suggested that more aggressive local therapy is beneficial to these patients,[6] CyberKnife radiosurgery might be used as an adjunct in the management of these lesions. There are several potential advantages to using radiosurgery in cases of PNS: reirradiation within previously radiated fields is possible; the high dose per fraction offers a better chance of tumor control than standard fractionation; and extremely conformal dose distributions can be achieved. This last advantage holds particularly true for CyberKnife radiosurgery, in which beams are distributed nonisocentrically and can thus conform better to target volumes with highly irregular shapes – which certainly describes the involved areas in patients with PNS.

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Development of a symptomatic intracranial meningioma in a male-to-female transsexual after initiation of hormone therapy

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ABSTRACT

Transsexual patients are routinely treated with sex hormone therapy, which may be a risk factor for meningiomas, although the data are largely inconclusive. Here we describe a male-to-female transsexual patient treated with estradiol patches, who developed an occipital meningioma causing generalized seizures. This is the second report of a male-to-female transsexual patient who developed a symptomatic meningioma after sex hormone treatment, adding to the growing evidence that sex hormones contribute to the pathogenesis of meningiomas. Meningiomas may therefore complicate hormone therapy and sex hormones may be contraindicated in transsexual candidates with meningiomas identified upon screening.

1. Introduction

Because meningiomas are twice as common in women than men,[1] sex hormones are likely to be involved in their pathogenesis. Transsexual patients, routinely given sex hormone doses several times higher than hormone replacement therapy (HRT), may have increased meningioma risk.[2,3] Male-to-female (MTF) sex hormone treatment (SHT) increases the risk of osteoporosis, cardiovascular disease,[2] and hormone-dependent tumors, including lactotroph adenomas, breast and prostate carcinoma.[2,4–6] We found one report of meningioma after SHT.[7] This is another case of symptomatic meningioma after SHT in a MTF transsexual.

2. Case report

A 36-year-old MTF transsexual, treated with estradiol 0.1 mg biweekly patches for more than 10 years, presented with headache and seizures. Imaging revealed a tentorial 4.75 × 4.0 × 3.7 cm left occipital extra-axial mass, not present on CT scan 1 month before SHT (Fig. 1).

The tumor was resected without complication. Histopathology revealed meningioma, strongly progesterone receptor-positive and estrogen-receptor negative (Fig. 2). After discharge on the 5th postoperative day with a normal neurological exam, the patient continued estradiol despite knowing the possible association of SHT and meningioma, but remained asymptomatic without recurrence for 5 years.

3. Discussion

This is the second report of a MTF transsexual individual developing intracranial meningioma after SHT. Whereas pituitary adenomas contraindicate SHT and prolactin levels are routinely followed in MTF transsexuals,[8] the role of SHT in meningiomas is poorly understood. Beyond the female predominance, evidence supporting a pathogenic role for sex hormones in meningioma is inconsistent: meningioma risk is increased or decreased with

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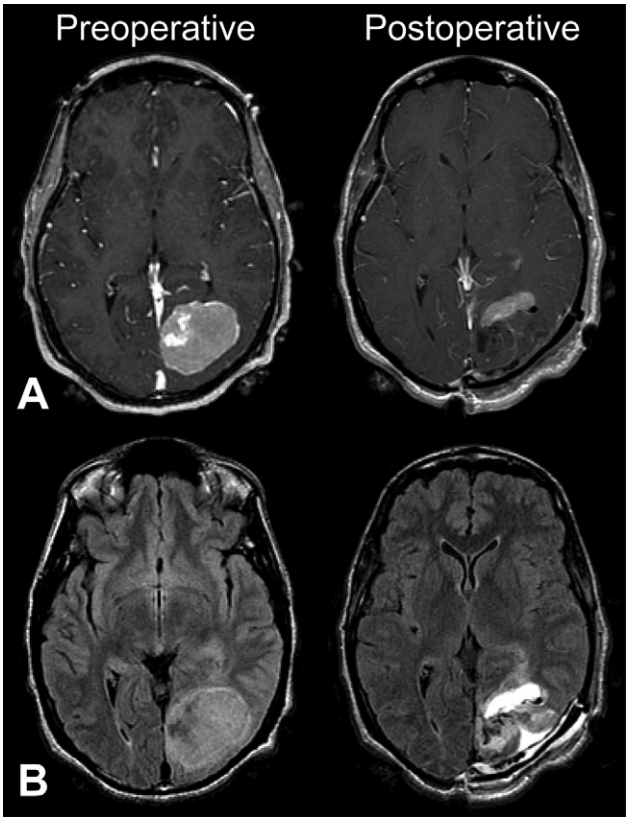

Fig. 1. (Left) Preoperative axial MRI showing a 4.75 × 4.0 × 3.7 cm left occipital extra-axial mass associated with the tentorium. (Right) Postoperative imaging 3 days after the operation was consistent with left occipital craniotomy with gross total resection of the enhancing portion of the tumor. (A) T1-weighted contrast enhanced MRI, (B) fluid attenuated inversion recovery MRI.

HRT and oral contraceptives in women, and with multiparity, obesity, and breast cancer.[1,9–16]

Most meningiomas express functional progesterone, not estrogen, receptors,[17,1,14] grow preferentially during the progesterone-predominant luteal phase,[15] and are more common in patients with lymphangioleiomyomatosis using progesterone agents.[18] There is one report of a patient with a meningioma arising after Norplant (progesterone agonist) placement.[19] The antiprogestin, mifepristone, inhibits meningioma cell growth *in vitro*,[20] and may induce minor regression of inoperable tumors, particularly in men and premenopausal women.[21] However, the only prospective mifepristone trial was equivocal.[17]

This evidence suggests that progesterone may contribute to meningioma pathogenesis, whereas trials of antiestrogens in recurrent or inoperable meningiomas have failed.[17] Interestingly, most meningiomas express somatostatin receptors, and somatostatin may retard tumor progression.[22,23]

Given the possible role of progesterone in the growth of meningiomas, it is concerning that MTF transsexuals are commonly treated with progestins.[2,3] The meningioma in this case may not be related to SHT. However, because meningioma prevalence in US men is at most 5:100,000, and the prevalence of MTF transsexuals is at most 1:10,000,[24,25] the probability of an incidental meningioma in a MTF transsexual is therefore about 1:200,000,000, suggesting that the association is not coincidental. Further study of the extent to which SHT increases the risk of meningioma is warranted. For patients currently on SHT and found to have a meningioma, cessation of hormones, particularly progestins, may be indicated.

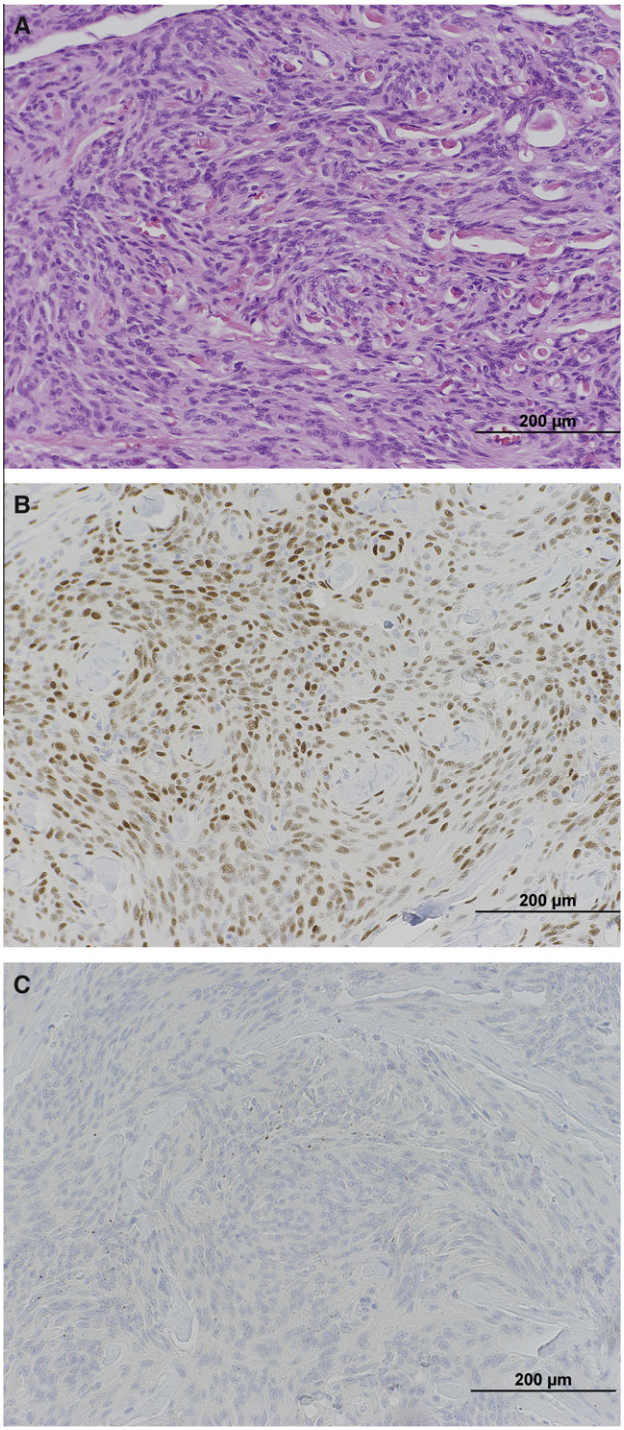

Fig. 2. Histopathologic staining of the tumor consistent with meningioma, World Health Organization grade 1, (A, haematoxylin and eosin) showing strong nuclear positivity with antibodies against the progesterone receptor (B), and no nuclear positivity with antibodies against the estrogen receptor (C).

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A rosette-forming glioneuronal tumour of the pineal gland

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ABSTRACT

A rosette-forming glioneuronal tumour (RGNT) is a rare tumour with new information emerging. We review the literature and describe the second patient with a RGNT arising from the pineal gland. This is also the fifth report of a RGNT arising from outside the fourth ventricle. The prognosis of RGNT remains guarded, as long-term follow-up is not yet available for most patients reported. With only one of 35 patients having a documented recurrence at 10 years, the prognosis seems favourable with appropriate, but not necessarily total, surgical resection.

1. Introduction

The rosette-forming glioneuronal tumour (RGNT) was initially described in 1998 as a midline infratentorial tumour, involving the fourth ventricle with both a neurocytic and a pilocytic astrocytoma component.[1] There have been 35 patients reported.[2–17] Only four patients have been reported with a RGNT arising outside the fourth ventricle. We report a patient with a RGNT arising from the pineal gland.

2. Case report

A 29-year-old man presented with 24 hours of progressive frontal headache, vomiting and decreasing level of consciousness. On examination he was drowsy but rousable (Glasgow Coma Scale score 13/15). His CT scan of the brain showed ventriculomegaly due to a hypodense heterogeneous lesion in the pineal gland. An MRI confirmed a $1.8 \times 2.0 \times 2.3$ cm mass with minimal contrast enhancement (Fig. 1). An external ventricular drain was inserted and the patient clinically improved.

The patient underwent a stereotactically guided suboccipital transtentorial craniotomy. A green–grey tumour was encountered in the pineal region with extension into the aqueduct. The tumour was removed and complete macroscopic resection was achieved.

Histopathology showed a glioneuronal tumour containing two distinct components: a glial component resembling pilocytic astrocytoma, and a neuronal component. In some areas these formed

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